

Posters

10. Gastroenterology/Liver disease/Metabolic complications of CF/Nutrition/Growth

S99

204 Decreasing fat-soluble vitamin deficiencies by increased awareness and change of vitamin preparation

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Objectives: We prescribe AquADEKs liquid, chewables or softgels in the recommended doses. This study compares the current status of plasma levels of A, D and E vitamins in all patients with our first assessment in 2010, where AquADEKs chewables were not available.

Methods: Plasma levels of A, D and E vitamins in adult and paediatric CF patients were obtained at annual assessment since 2009. Deficiency was described as below lower level of normal local values (vit D 50nmol/l; vit A 0.7µmol/l for <7y, 0.91 for ≥7y; vit E 17µmol/l). In case of deficiency the cause was discussed with patients.

Results: The mean value for vitamin D in summer was 79 and in winter 70 with no significant difference ($p=0.062$). The results for vitamin A and E deficiency are comparable to the vitamin D as shown in Table 1. In the age group 12–17 years the vitamin A deficiency has decreased from 37.5% in the first sample to 10.3% in the recent sample and vitamin E deficiency has decreased from 68.8% to 30%. At first sample 54.4% of patients had at least one deficiency, reduced to 29.6% in recent sample.

Table 1. Vitamin D deficiency by age group

Age group	First sample		Most recent sample	
	Patients in age group	Patient(s) with vit D deficiency	Patients in age group	Patient(s) with vit D deficiency
0–5 years	40	1 (2.5%)	13	0 (0%)
6–11 years	25	3 (12%)	35	1 (2.9%)
12–17 years	33	16 (48.5%)	30	7 (23.3%)
18+ years	52	20 (38.5%)	68	17 (25%)

Conclusion: We found a significant reduction in the number of patients with fat-soluble vitamin deficiency as a result of common effort and awareness in the team. Especially, vitamin A and E has shown more than a 50% reduction in deficiency. For most patients the AquADEKs chewables are more palatable, which may result in better adherence.

205 Folate deficiency in adult patients at a regional cystic fibrosis (CF) centre

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Water soluble vitamin deficiency is not thought to be a problem in patients with CF. We describe serum folate levels (SFL) in an adult CF population and characterise those needing SFL monitoring.

Methods: Retrospective review of electronic patient records (2007 to date) was used to extract initial SFL. SFL was compared by gender, PPI use in year pre SFL, liver disease, pancreatic and transplant status. Correlation of SFL to Hb, B12, FEV1% predicted (FEV1), BMI and IV antibiotic use in year pre SFL (IVs) was calculated. Optimal (5.4–24 µg/l) and suboptimal (<5.4µg/l) SFL groups were compared.

Results: 211 patients (98M/113F) had a SFL; 28.4% had sub-optimal levels. Hb differed significantly by SFL group, [median (IQR)] [116 (35) vs 132 (39), $p=0.009$] but BMI [20.99 (3.99) vs 21.22 (4.57), $p=0.537$], FEV1 [56 (34.25) vs 59 (40.75), $p=0.274$], IVs [28 (69) vs 16 (55), $p=0.174$] and B12 [432 (247) vs 498 (338), $p=0.125$] did not. Correlations for SFL vs Hb ($p=0.01$) and SFL vs B12 ($p=0.05$) were positive and significant. No significant correlation was found for FEV1 (positive, $p=0.158$), BMI (positive, $p=0.619$) or IVs (negative, $p=0.132$). SFL differed significantly by gender [M 8.3 (6.1) vs F 7.0 (6.3), $p=0.043$] but not by pancreatic status [PS 5.0 (5.4) vs PI 7.7 (6.3), $p=0.060$], transplant status [pre 7.6 (6.1) vs post 7.3 (8.2), $p=0.898$], liver disease [LD 7.4 (5.9) vs no LD 7.6 (6.6), $p=0.688$] or PPI use [PPI 7.8 (6.6) vs no PPI 7.4 (6.2), $p=0.171$].

Conclusion: Folate deficiency exists in adults with CF. Low Hb, B12 and female gender may indicate a need to monitor SFL. Further exploration of SFL looking at genetics, gastric surgery, types of antibiotics, zinc and MCV may be useful.

206 Serum zinc concentration in cystic fibrosis patients with CFTR I1234V mutation associated with pancreatic sufficiency

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Objective: Zinc (Zn) has a wide range of physiologic functions, and related to pulmonary health, immunity, and growth. Zn deficiency has been observed in a subset of patients with cystic fibrosis (CF). The aim of the study was to determine serum Zn level among CF patients with CFTR I1234V mutation associated with pancreatic sufficiency (PS).

Methods: Cross-sectional study of CF patients. Data on age, weight, height, body mass index (BMI), BMI Z-score, FEV1 and chronic *Pseudomonas aeruginosa* infection were collected. Serum Zn, albumin and total proteins were measured.

Results: Fifty-three CF patients (31 females) were enrolled in this study, 45 CF patients with CFTR I1234V mutation. The remaining 8 CF patients with other mutations associated with pancreatic insufficiency (PI). Patients ranged from 2 to 49 years with a mean age of 15.1 ± 9.1 years with mean plasma Zn of 0.78 ± 0.15 mcg/ml. Seven (13.2%) patients with CFTR I1234V and PS had low Zn levels, below 0.6 mcg/ml, when standard reference values were used for classification. Mean age in the Zn deficient group was significantly higher, 23.0 ± 12.2 years compared to that observed in the normal Zn levels group, 13.9 ± 8.1 years ($p=0.013$). The mean FEV1 (% predicted) in the deficient group was found to be lower, $66.5 \pm 29.6\%$ compared to patients with normal Zn levels, $81.4 \pm 20.4\%$ ($p=0.043$). Persistent *P. aeruginosa* colonization was more prevalent in Zn deficient group. BMI Z-score positively associated with Zn levels.

Conclusion: These findings suggest that Zn deficiency can occur in patients with CF and PS. The association of Zn levels and the frequency of *P. aeruginosa* isolated in CF patients need further investigation.

207 Cow's milk allergy in cystic fibrosis children

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Objectives: In CF an important feature is the pancreatic insufficiency expressed by steatorrhea; in some cases, despite enzyme supplementation the chronic diarrhea is poorly controlled.

Aim of study: To find the prevalence of cow's milk allergy among patients with cystic fibrosis.

Methods: In one year, sixty-seven children with cystic fibrosis, aged 1 month to 3 years (19 infants), followed in our center were observed for CMPA. Cow's milk allergy was suspected in the presence of specific clinical manifestation (gastrointestinal symptoms, cutaneous signs, respiratory features), in cases with a suggestive medical history (failure to thrive, colics etc). In addition CMPA-specific IgE (α and β lactoglobulin, casein) and diagnostic elimination test + food challenge test.

Results: Among infants, CMPA was diagnosed in 47.36% (9 patients) of children, predominantly in pancreatic insufficient children (77% – CF infant with pancreatic insufficiency). Toddlers (1–3 years) were diagnosed in a smaller percent, only 16.6% (8 children) proved to have CMPA, although in more than 35% of toddlers, a positive history for CMPA diagnosis was found. Cystic fibrosis patients with pancreatic insufficiency associated more frequently CMPA (88.23%) than cystic fibrosis patients pancreatic sufficient. The prevalence of cow's milk allergy was important, cumulating a 25.37% of CF patients.

Conclusion: Cow's milk allergy was frequently found in CF children, especially associated with pancreatic insufficiency. The “combined” enteropathy could influence the disease's outcome and should be considered especially in persistent diarrhea of CF children with correct enzyme supplementation.